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Congenital left atrial wall aneurysm in a patient with neurofibromatosis

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SUMMARY A congenital intrapericardial aneurysmal dilatation of the left atrial wall was found in a 28 year old man who presented with atrial fibrillation after a syncopal event. The patient had cutaneous manifestations of neurofibromatosis. The diagnosis was made by cross sectional echocardiography and confirmed by angiocardiography. Surgical excision of the aneurysm resolved the symptoms.

Congenital intrapericardial aneurysm of the left atrium is a rare anomaly. There are 32 reports of aneurysms affecting the atrial appendage alone[1–8] and 13 reports of aneurysms of the atrial wall.[9–11] We report a case in which there was an intrapericardial aneurysm of the left atrial wall and we review the mode of presentation, investigation, potential complications, and possible association with neurofibromatosis.

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Case report

A 28 year old man presented to a local hospital after a syncopal episode associated with tight central chest pain and severe dyspnoea while playing football. On

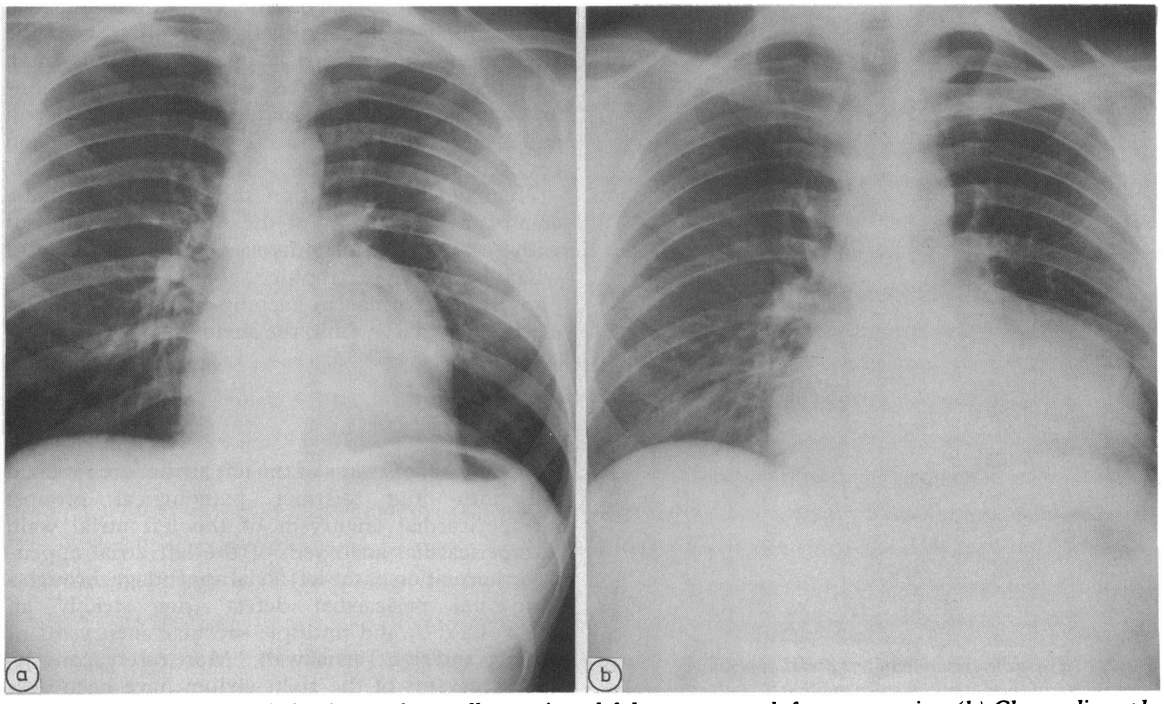

Fig 1 *(a) Chest radiograph showing an abnormally prominent left heart ten years before presentation. (b) Chest radiograph showing cardiomegaly at presentation.*

arrival he was conscious but hypotensive, and electrocardiographic monitoring showed atrial flutter with variable block. When the arrhythmia was found to be resistant to cardioversion and intravenous amiodarone he was transferred to the regional cardiothoracic centre.

He reported increasing exertional dyspnoea over the past two months. There was no recent history of viral illness or a family history of cardiovascular disease.

He was clinically in atrial fibrillation with a ventricular rate of 160 per minute but there was adequate perfusion with a blood pressure of 100/60 mm Hg. A pulsation was seen in the pulmonary area and the apex was not displaced. The jugular venous pulse was not elevated. Heart sounds were normal with a soft ejection systolic murmur at the left sternal edge. Lung fields were clear. He had seven café au lait spots and bilateral axillary freckling.

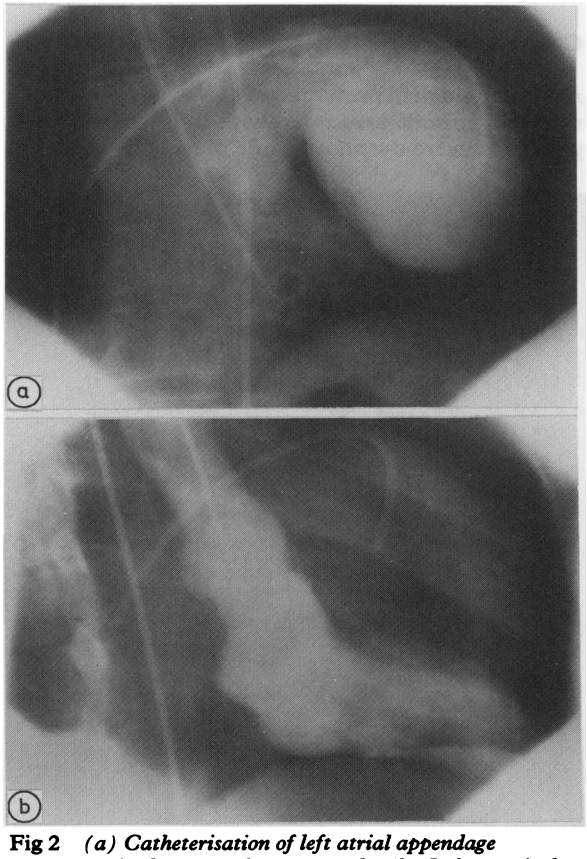

Fig 2 *(a) Catheterisation of left atrial appendage aneurysm via the patent foramen ovale. (b) Left ventricular angiogram showing compression of left ventricle by left atrial aneurysm.*

The 12 lead electrocardiogram confirmed atrial fibrillation and showed right axis deviation, partial right bundle branch block, poor R wave progression, and deep S waves in V2 and V3. A chest radiograph obtained 10 years before showed an abnormality of the mid left heart border (fig 1a). The chest radiograph at presentation showed considerable cardiomegaly with a cardiothoracic ratio of 19:30 (fig 1b). Cross sectional echocardiography showed normal left ventricular size and function. Adjacent to and impinging on the left ventricle was a large echo-free chamber. Doppler examination confirmed flow within the chamber and colour Doppler suggested that it communicated with the left atrium close to the mitral valve. Cardioversion with energies up to 400 J was attempted but failed. Treatment with digoxin improved the control of ventricular rate and symptoms.

Right heart catheterisation showed normal pressures and no intracardiac shunt but the left atrium and the additional chamber were entered through a patient foramen ovale. Injection of contrast confirmed communication between the chamber and the left atrium (fig 2a), and the left ventricular angiogram suggested compression of the left ventricle during diastole, presumably by the large aneurysm (fig 2b).

A dilated aneurysm of the left atrial wall with a maximum diameter of 8 cm was found at operation through a left thoracotomy. Electrophysiological studies showed electrical activity in all areas of the aneurysm. The left atrial appendage was normal. The aneurysm was resected without cardiopulmonary bypass. Post-cardiotomy syndrome developed and lasted three days and subsequent cardioversion restored sinus rhythm.

Histological examination showed fibrosis in the aneurysm wall mainly of the subendocardium but with residual irregular myocardial fibres, many of which were hypertrophied. Neural fibres were present and normal, in keeping with the finding of electrical activity within the aneurysm.

Discussion

Congenital aneurysms of the left atrium are rare and fall into four distinct pathological groups: intrapericardial aneurysm of the left atrial wall; intrapericardial aneurysm of the left atrial appendage; herniation of the left atrial appendage through a congenital pericardial defect (not strictly an aneurysm)[12-14]; and multiple saccular aneurysms of the left (and right) atrial wall.[15] More rarely, congenital aneurysms of the right atrium have been described[16 17]; in one case there was also an aneurysm of the left atrial wall.[18]

Table *A comparison of reported findings in aneurysms of the left atrial wall and appendage*

Patient characteristics	*Left atrial*	
	Wall aneurysm No (%)	*Appendage aneurysm No (%)*
Case reports	14 (100)	32 (100)
Clinical features:		
Palpitation/SVT	10 (71)	16 (50)
Chest pain	2 (14)	4 (13)
Systolic murmur	8 (57)	10 (31)
Systemic embolism	0 (0)	8 (25)
Electrocardiogram:		
Atrial fibrillation/SVT	4 (29)	9 (28)
Chest radiograph:		
Mid left heart bulge	3 (21)	22 (69)
Cardiomegaly	11 (79)	10 (31)
Operative findings:		
Mural thrombus	1 (7)	4 (13)

SVT, sustained ventricular tachycardia.

The table summarises findings reported in aneurysms of the left atrial wall and appendage. Aneurysms are equally common in male and female patients aged 1–69 years. Aneurysms of the left atrial wall usually arise from the posterior wall and may extend to the mitral ring. There are three reports of the aneurysm affecting the entire left atrium.[9 18] Aneurysms of the left atrial appendage usually have a narrow neck and are separate from a normal sized left atrium. As in our patient, paroxysmal or sustained atrial arrhythmia is the usual mode of presentation. A routine chest radiograph is often the first indication of cardiac disease in a symptom free patient. Occasionally, the condition presents as atypical chest pain. Intraventricular conduction defects and T wave abnormalities have been described.[8] The chest radiograph is always abnormal (table). A discrete bulge in the mid left heart in a symptom free patient is also the usual presentation when there is herniation of the left atrial appendage,[13] but there are isolated reports of chest pain, pericarditis, and strangulation of the appendage.[14]

Clinical differentiation between the aneurysms may be difficult. Cross sectional echocardiography may show congenital pericardial defects[8 12] and exclude extracardiac causes for the abnormal cardiac shadow. In this case combined echocardiographic and Doppler examination confirmed communication of the aneurysm with the left atrium but we were unable to distinguish between an aneurysm of the atrial wall or the atrial appendage. Radionuclide blood pool scanning[5] and computed tomography may be useful. Cardiac catheterisation with angiography has been most widely used to establish a definitive diagnosis.

The major risk of an aneurysm of the left atrial appendage is systemic (usually cerebral) embolism (table). None of the 13 patients with aneurysms of the atrial wall had systemic emboli, although at operation one had a thrombus in the aneurysm. It seems advisable to recommend anticoagulants in both groups at diagnosis in view of the risk of cerebral embolism.

It is not surprising that atrial arrhythmias are common in view of the fibrosis, hypertrophy, and dilatation of the atrial wall. As with previous reports, restoration of sinus rhythm was difficult and was only possible after the aneurysm had been resected. Operation is the recommended treatment because it also abolishes symptoms and removes the source of emboli.[1] Operation is recommended for extrapericardial herniations because of the risk of strangulation.[12] To date there has been no reported mortality related to operation.

The assertion that intrapericardial aneurysms are congenital is based on pathological examination and the association with other cardiac anomalies.[11] The aneurysmal dilatation is thought to be the result of congenital weakness in the wall. Hypertrophy of the atrial myocardium with a thickened endocardium or the development of fibroelastosis has been reported.[10]

To our knowledge, this is the first reported case of left atrial aneurysm and neurofibromatosis. The presence of seven café au lait spots and bilateral axillary freckling is pathognomic of neurofibromatosis. There was no family history of the disease; however, new mutations account for 50% of all index cases. Close clinical examination did not find any other manifestations of the disease. In one series 7·7% of children with neurofibromatosis had congenital heart disease.[19] A strong association between neurofibromatosis and pulmonary valve stenosis has been described.[20] Large series have suggested that the association between neurofibromatosis and congenital heart disease is coincidental. Because neurofibromatosis is often not diagnosed until the third decade, however, a true association might have been overlooked.

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